2.10 Systemic Diseases and the Kidney

**FIGURE 2-22**
Approximate relative frequency of P-ANCA/M PO-ANCA versus C-ANCA/PR3-ANCA in patients with pauci-immune necrotizing and crescentic glomerulonephritis without systemic vasculitis (“renal-limited vasculitis”), microscopic polyangiitis, and Wegener’s granulomatosis. Note that most patients with renal-limited disease have P-ANCA/M PO-ANCA, most patients with Wegener’s granulomatosis have C-ANCA/PR3-ANCA, and patients with microscopic polyangiitis do not have a major preponderance of either ANCA specificity.

**FIGURE 2-23** (see Color Plate)
Early segmental fibrinoid necrosis and infiltration by neutrophils in an ANCA-positive patient with Wegener’s granulomatosis (Masson trichrome stain). There also is fibrin (red/fuchsinophilic material) in Bowman’s space, which is a precursor event to crescent formation.

**FIGURE 2-24**
Glomerulus from a patient with ANCA and a pauci-immune necrotizing and crescentic glomerulonephritis showing a large circumferential crescent and segmental lysis of glomerular basement membranes (combined Jones silver and hematoxylin and eosin stain). Also note the adjacent tubulointerstitial inflammation, which often is pronounced in ANCA disease. This pattern of glomerular injury can be seen with any of the ANCA-small vessel vasculitides.

**FIGURE 2-25** (see Color Plate)
Direct immunofluorescence microscopy demonstrating intense staining of a crescent and adjacent segmental glomerular fibrinoid necrosis with an antiserum specific for fibrin in a renal biopsy from a patient with ANCA small vessel vasculitis. There was no staining of glomeruli in this specimen with antisera specific for IgG, IgA, or IgM.
2.11 Vasculitis (Polyarteritis Nodosa, Microscopic Polyangiitis, Wegener’s Granulomatosis, Henoch-Schönlein Purpura)

**FIGURE 2-26**
Chronic ANCA-associate glomerulonephritis with effacement of the architecture of a glomerulus by extensive sclerosis. Bowman’s capsule has been destroyed and there is periglomerular fibrosis and chronic inflammation.

**FIGURE 2-27**
Necrotizing arteritis involving an interlobular artery in a renal biopsy specimen from a patient with ANCA-positive microscopic polyangiitis (hematoxylin and eosin). There is focal transmural fibrinoid necrosis with intense perivascular inflammation. This pattern of arteritis is nonspecific, and could be seen, for example, in a patient with polyarteritis nodosa, microscopic polyangiitis, or Wegener’s granulomatosis. The presence of ANCA or glomerulonephritis in the patient would exclude polyarteritis nodosa.

**FIGURE 2-28**
Direct immunofluorescence microscopy demonstrating intense staining of the fibrinoid necrosis in the wall of an interlobular artery with an antiserum specific for fibrin in a renal biopsy from a patient with microscopic polyangiitis.

**FIGURE 2-29**
Medullary leukocytoclastic angiitis involving vasa recta in a patient with Wegener’s granulomatosis (hematoxylin and eosin). When this process is severe, papillary necrosis may result. The frequency of this process is unknown because the medulla often is not sampled in renal biopsy specimens.
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**FIGURE 2-30**
Poorly defined focus of necrotizing granulomatous inflammation in the cortex in a renal biopsy obtained from a patient with ANCA-positive Wegener's granulomatosis (hematoxylin and eosin). Granulomatous inflammation is only very rarely observed in renal biopsy specimens.

**FIGURE 2-31**
Necrotizing granulomatous inflammation in a wedge biopsy of lung from a patient with Wegener's granulomatosis (hematoxylin and eosin). Note the scattered large multinucleated giant cells on the left side and the extensive necrosis and neutrophilic infiltration on the right side. The granulomatous inflammation of acute Wegener's granulomatosis has much more neutrophilic infiltration and liquefactive necrosis than most other forms of granulomatous inflammation, which is why the lesions in the lung tend to cavitate, and why the lesions in the nose and sinuses tend to destroy bone.

**FIGURE 2-32**
Hemorrhagic alveolar capillaritis in a wedge biopsy from the lung of a patient with microscopic polyangiitis (hematoxylin and eosin). Note the neutrophils within alveolar capillaries and the massive hemorrhage into the air spaces. This pattern of injury can be seen in both microscopic polyangiitis and Wegener's granulomatosis. The pulmonary hemorrhage of anti-GBM disease usually does not have conspicuous neutrophils in alveolar capillaries.

**FIGURE 2-33**
Categorization of patients with crescentic glomerulonephritis with respect to both the immunopathologic category of disease (immune complex versus anti-GBM versus ANCA) and the clinicopathologic expression (glomerulonephritis alone versus Wegener's granulomatosis versus Goodpasture's syndrome versus other small vessel vasculitis) [11]. Note that most patients with ANCA have some expression of systemic vasculitis rather than glomerulonephritis alone. Most patients with Wegener's granulomatosis have C-ANCA/PR3-ANCA but some have P-ANCA/MPO-ANCA. Also note that some patients with anti-GBM and some patients with immune complex disease also are ANCA positive. (Adapted from Jannette [11]).
References